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Case report

Idiopathic renal replacement lipomatosis

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Key words

Renal replacement lipomatosis

Summary

Renal replacement lipomatosis (RRL) is a rare disorder which exhibits extensive proliferation of fatty tissue within the renal sinus, hilum, and perirenal region. The pathogenesis of this entity is unknown, though association with aging, renal atrophy, long-standing chronic urinary infections has been noted. Although imaging modalities may suggest the diagnosis of this entity, it is histopathology that clinches the diagnosis most accurately. We report a case of a 52 year old male who presented with nonfunctioning kidney and was histopathologically confirmed to be a case of renal replacement lipomatosis.

Introduction

Renal replacement lipomatosis (RRL) is a rare benign condition which exhibits proliferation of renal sinus/ hilar and perirenal adipose tissue associated with marked atrophy of the renal parenchyma [1]. Nearly seventy percent cases are related to renal calculi with chronic inflammation and hydronephrosis [2]. This entity may radiologically and clinically mimic a fatty neoplasm or xanthogranulomatous pyelonephritis of kidney.

Case report

A 52 year old male was admitted to our hospital with complaint of continuous and dull aching right flank pain since four months. Physical examination was largely unremarkable. Renal ultrasound abdomen revealed an oval echogenic mass 5.8 x 2.3 cm in size with loss of cortico-medullary differentiation in right renal fossa. The contralateral kidney showed compensatory hypertrophy with size being 11.5 x 5.6 cm. There was no calculus or hydronephrosis. Thus, possibility of medical renal disease leading to small shrunken kidney was kept. The other possibility kept was that of a dysplastic kidney or a renal artery stenosis. MR urogram revealed a small, shrunken right kidney and bilaterally normal pelvicalyceal system. Further, renal DTPA scan revealed a small, contracted poorly functional right kidney with GFR of only 4.0 ml/minute (Fig. 1a). Since the right kidney was deemed nonfunctional, right nephrectomy was performed.

Gross examination showed a distorted nephrectomy specimen 5 x 3.5 x 2.5 cm with intact Gerota's fascia. On cut section, corticomedullary differentiation appeared to be lost. Most of the renal parenchyma was replaced with adipose tissue with a thin rim of atrophied renal parenchyma at the periphery (Fig. 1b).

Microscopic examination showed extensive fatty infiltration in the renal parenchyma with clear demarcation between the adipose tissue and the residual renal parenchyma (Fig. 2a). The residual renal parenchyma at the periphery showed marked atrophy in the form of sclerosed glomeruli, thyroidisation of tubules, thickened blood vessels and interstitial fibrosis (Fig. 2b). Thus, a diagnosis of renal replacement lipomatosis of the right kidney was made.

Discussion

RRL is an unusual entity that was first described by Kutzmann in 1931 [3]. It represents a spectrum of changes and terms such as 'renal sinus lipomatosis', 'replacement lipomatosis', and 'fibrolipomatosis of kidney' have been used for this rare condition [4]. It is considered to be a degenerative process, in contrast to renal lipomas which are neoplastic. The mildest form of this disease

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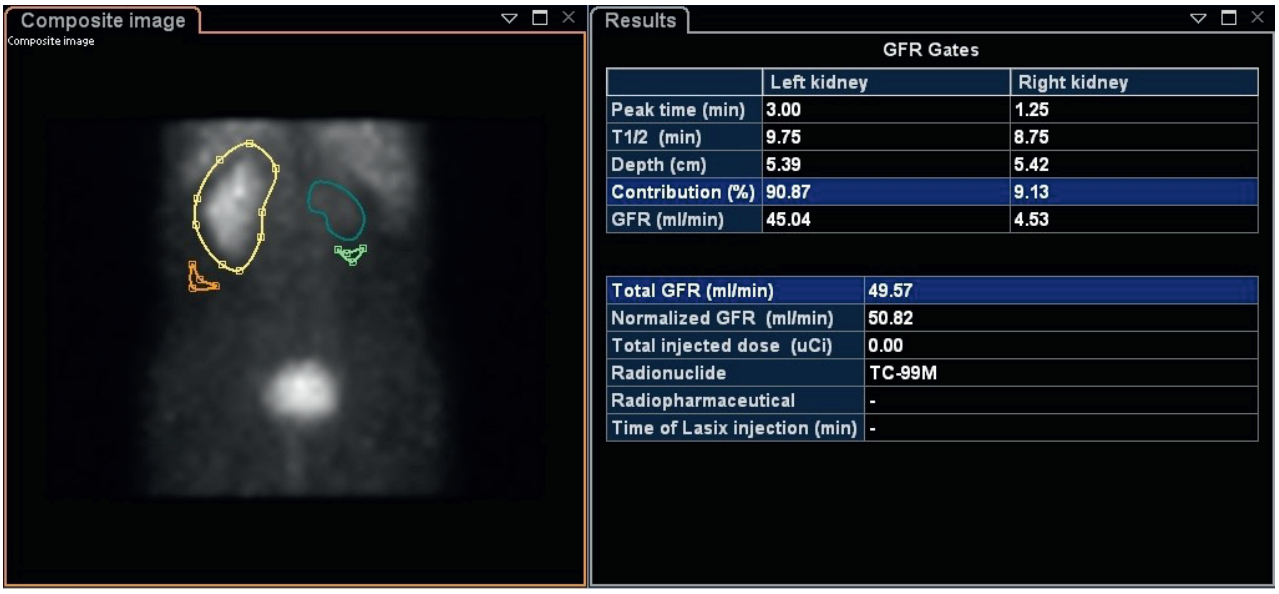
Fig. 1a. Renal DTPA scan showing shrunken right kidney with marked reduction of GFR.



	Left kidney	Right kidney
Peak time (min)	3.00	1.25
T1/2 (min)	9.75	8.75
Depth (cm)	5.39	5.42
Contribution (%)	90.87	9.13
GFR (ml/min)	45.04	4.53

Total GFR (ml/min)	49.57
Normalized GFR (ml/min)	50.82
Total injected dose (uCi)	0.00
Radionuclide	TC-99M
Radiopharmaceutical	-
Time of Lasix injection (min)	-

Fig. 1b. Nephrectomy specimen showing distorted shrunken kidney replaced by adipose tissue and marked loss of renal parenchyma.

Fig. 2a. Fatty infiltration of residual renal parenchyma showing marked atrophy (H&E, x 40).

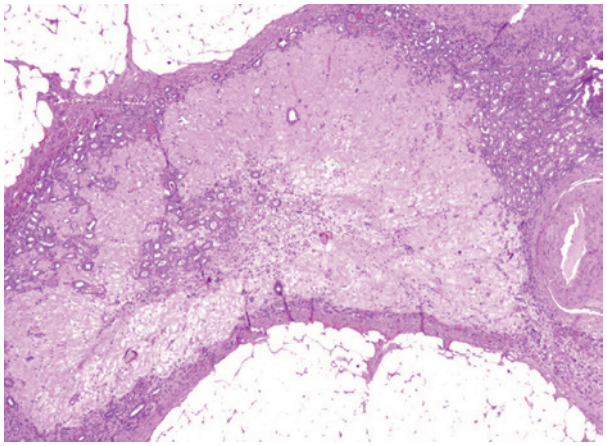
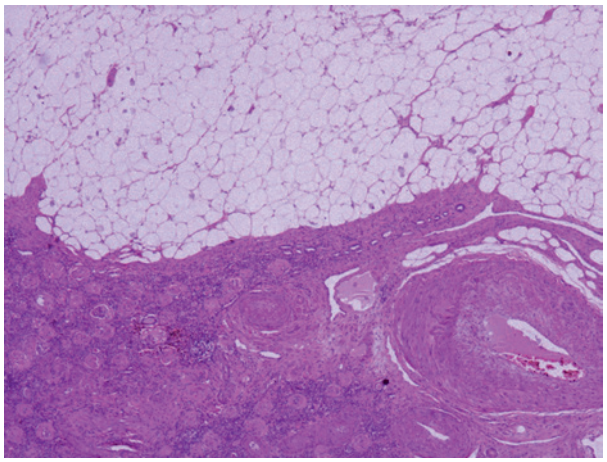


Fig. 2b. Clear demarcation of adipose tissue with atrophied renal parenchyma showing sclerosed glomeruli and thick walled blood vessels (H&E, x 200).



is renal sinus lipomatosis, which is usually seen in the sixth and seventh decade and is associated with obesity, atherosclerosis or use of exogenous steroids and occasionally post renal transplant; a process not specific to the kidney but seen in any organ which atrophies, such as the thymus or senile pancreas [4-6]. However, severe loss of renal parenchyma with massive fat deposition is commonly associated with long standing inflammation and calculi [2].

Replacement lipomatosis is usually unilateral in occurrence and rarely idiopathic with the cause being identified in majority of cases. The present case was rarest of rare and was of idiopathic variety as none of the aforementioned etiological factors were detected in the past or present either clinically or radiologically. The age group commonly affected is fourth to sixth decade as was seen in the our case, though it has also been reported in a young patient in second decade with posterior mediastinal lipomatosis [7]. Clinical symptoms include urinary tract infections, fever, and flank pain and occasionally obstruction, due to mass effect exerted by the increases proliferation in the renal sinus on the intrarenal collecting structures [8]. Blood urea nitrogen and serum creatinine is usually within the normal limits, though in our case, it was elevated. They are normal if the disease is unilateral with other well functioning kidney.

Ultrasonography may suggest the diagnosis by demonstrating parenchymal atrophy or a hyperechoic renal sinus mass with a stone; however, it is not diagnostic. CT scan can confirm the fatty infiltration in the kidney and its distribution. Malakoplakia, xanthogranulomatous pyelonephritis, fat-containing neoplasms including lipoma, liposarcoma, and angiomyolipoma and transitional carcinoma of the renal sinus are the usual differential diagnosis reported in the literature with imaging techniques [9]. Associated findings like renal atrophy, calculus, and hydronephrosis can exclude the possibility of lipomatous tumors [10]. Xanthogranulomatous pyelonephritis can be extremely difficult to differentiate from RRL as both entities show overlapping clinicoetiologic features. However, chronic obstruction in XP causes destruction of renal parenchyma, and histological examination reveals lipid-laden macrophages (foamy histiocytes) diffusely infiltrating the renal parenchyma, in contrast to RRL where fat cells remain outside of the atrophied renal parenchyma.

Grossly the kidney can be enlarged or small in size with markedly atrophic renal parenchyma as was seen in our case [3 8]. The reniform shape of the kidney is always preserved. Histologically, this condition shows mature adipose tissue replacing the renal parenchyma leading to its atrophy. There can be sharp demarcation between the adipose tissue and the preserved renal parenchyma as seen in this case [3]. This process is distinct from that seen with lipomas, which are found within the parenchyma.

To conclude, RRL is a relatively uncommon entity and may be confused with renal neoplasms on radiology. It usually presents with a poorly or non-functioning kidney and histologically shows replacement of the renal parenchyma with mature adipose tissue leading to end stage renal disease. This report is an attempt to increase the awareness of this entity and its histopathological features to avoid any misdiagnosis.

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